

Kawasaki Syndrome

1) THE DISEASE AND ITS EPIDEMIOLOGY

A. Etiologic Agent

The cause of Kawasaki syndrome is unknown but is presumably an infectious or toxic agent.

B. Clinical Description

Kawasaki syndrome is a multi-system disease primarily affecting children under 5 years old. It is an acute, self-limited, systemic vasculitis characterized by an initial high spiking fever that can persist for 1 to 2 weeks. It does not respond to antibiotics, and it is associated with extreme irritability and mood change. The fever is accompanied by bilateral conjunctivitis (seen in about 85% of cases); changes to the lips and mouth including dry, red or cracked lips, a sore red throat, and/or strawberry tongue (90%); changes to the extremities including peeling of skin, rashes and/or swelling of the hands and feet (75%); a generalized red rash affecting the trunk or perineal region (80%); and an enlarged cervical lymph node (usually solitary, often unilateral) (70%). Other common symptoms include severe abdominal cramping, diarrhea and vomiting.

The convalescent (recovery) phase can be lengthy. Cardiac involvement is a major cause of morbidity and mortality associated with Kawasaki syndrome. Approximately 20% of untreated cases develop coronary artery aneurysms within 6 weeks. Prompt treatment with intravenous immune globulin (IVIG) and aspirin can reduce aneurysm formation to about 8% of cases, as well as reducing fever and inflammatory signs. The case-fatality rate for Kawasaki syndrome is 0.1% to 1%. Other complications can involve any organ of the body.

C. Reservoirs

Reservoirs are unknown.

D. Modes of Transmission

Modes of transmission are unknown.

E. Incubation Period

The incubation period is unknown.

F. Period of Communicability or Infectious Period

The disease is not known to be communicable from person-to-person.

G. Epidemiology

Kawasaki syndrome occurs worldwide with most cases reported from Japan, where nationwide epidemics have been documented. The estimated number of new cases each year in the United States is approximately 2000. The peak age of occurrence in the United States is between 18 and 24 months, with 80% of cases reported in children younger than 5. Males are affected more frequently than females, and children of Asian descent have the highest incidence rate. Epidemics occur on a 2 to 3 year cycle, usually during the winter and spring. While the mode of transmission for Kawasaki syndrome has not been documented, siblings (especially twins) of cases are more likely to be affected than the general population.

2) REPORTING CRITERIA AND LABORATORY TESTING SERVICES

A. What to Report to the Massachusetts Department of Public Health

- A healthcare provider's clinical diagnosis of Kawasaki syndrome.

Note: See Section 3) C below for information on how to report a case.

B. Laboratory Testing Services Available

The laboratory features of Kawasaki syndrome are nonspecific and nondiagnostic; therefore no testing services are offered at the Massachusetts State Laboratory Institute.

3) DISEASE REPORTING AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

- To identify disease clusters and demographic characteristics.

B. Laboratory and Healthcare Provider Reporting Requirements

Refer to the lists of reportable diseases (at the end of this manual's Introduction) for information.

C. Local Board of Health Reporting and Follow-Up Responsibilities.

1. Reporting Requirements

Massachusetts Department of Public Health (MDPH) regulations (*105 CMR 300.000*) stipulate that each local board of health (LBOH) must report the occurrence of any case of Kawasaki syndrome, as defined by the reporting criteria in Section 2) A above. Current requirements are that cases be reported to the MDPH Division of Epidemiology and Immunization, Surveillance Program using an official CDC *Kawasaki Syndrome Case Reporting* form (in Appendix A). Refer to the *Local Board of Health Reporting Timeline* (at the end of this manual's introductory section) for information on prioritization and timeliness requirements of reporting and case investigation.

2. Case Investigation

- a. It is the LBOH responsibility to complete a CDC *Kawasaki Syndrome Case Reporting* form (in Appendix A) by interviewing the case and others who may be able to provide pertinent information. Much of the information required on the form can be obtained from the case's healthcare provider or the medical record.
- b. Use the following guidelines to assist you in completing the form:
 - 1) Accurately record the demographic information, date of symptom onset, whether hospitalized (and associated dates), outcome of disease, and whether the patient has recurrent Kawasaki syndrome. (Although the form asks only for the case's initials, please include the complete name and address. There is extra room at the bottom of the second page.)
 - 2) Collect the information requested in the "Diagnostic Criteria" section. This information is important in defining a case. You may ask the healthcare provider to submit a copy of the medical record to you or enlist his/her aid in completing these sections of the case report form.
 - 3) Collect as much information as possible about cardiac studies/results and complications associated with the illness.
 - 4) If you have made several attempts to obtain case information, but have been unsuccessful (*e.g.*, the case or healthcare provider does not return your calls or respond to a letter, or the case refuses to divulge information or is too ill to be interviewed), please fill out the form with as much information as you have gathered. Please note on the form the reason why it could not be filled out completely.
- c. After completing the form, mail (in an envelope marked "Confidential") to the MDPH Division of Epidemiology and Immunization, Surveillance Program. The mailing address is:
MDPH, Division of Epidemiology and Immunization
Surveillance Program, Room 241

305 South Street
Jamaica Plain, MA 02130

Note: Do not send the case report form to CDC as it indicates.

- d. Institution of disease control measures is an integral part of case investigation. It is the LBOH responsibility to understand, and, if necessary, institute the control guidelines listed below in Section 4), Controlling Further Spread.

4) CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements (105 CMR 300.200)

None.

B. Protection of Contacts of a Case

None.

C. Managing Special Situations

Reported Incidence Is Higher than Usual/Outbreak Suspected

If multiple cases of Kawasaki syndrome occur in your city/town, or if you suspect an outbreak, investigate clustered cases. Identify common factors, such age, school, workplace or activities to help elucidate risk factors. Consult with the Division of Epidemiology and Immunization at (617) 983-6800 or (888) 658-2850. The Division can help determine a course of action to prevent further cases and can perform surveillance for cases that may cross several town lines and therefore be difficult to identify at a local level.

D. Preventive Measures

None.

ADDITIONAL INFORMATION

There is no formal Centers for Disease Control and Prevention (CDC) surveillance case definition for Kawasaki syndrome. (CDC case definitions are used by the state health department and CDC to maintain uniform standards for national reporting.) Diagnostic decisions are made based on clinical presentations when there are no other reasonable explanations for the illness. For reporting a case to the MDPH, always refer to the criteria in Section 2) A.

REFERENCES

American Academy of Pediatrics. *Red Book 2000: Report of the Committee on Infectious Diseases*, 25th Edition. Illinois, American Academy of Pediatrics, 2000.

Chin, J., ed., *Control of Communicable Diseases Manual*, 17th Edition. Washington, DC, American Public Health Association, 2000.

Mandell, G., Bennett, J., Dolin, R., eds. *Principles and Practice of Infectious Diseases*, Fourth Edition. New York, Churchill Livingstone Inc., 1995.

MDPH. *Regulation 105 CMR 300.000: Reportable Diseases and Isolation and Quarantine Requirements*. MDPH, Promulgated November 1998, (Printed July 1999).